

## Emergency resection of obstructing thyroid lymphoma causing stridor: A case report

Emergency resection of an obstructing non-hodgkin thyroid lymphoma

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### Abstract

Primary thyroid lymphoma is a relatively rare histological entity, accounting for about 2% of extra-nodal lymphomas and only 0.5-5% of all thyroid malignancies. The incidence rate is just two per million individuals, best treatment options and critical prognostic variables have yet to be determined. Herein, we report the case of a 48-year-old woman who presented with acute stridor and life-threatening airway obstruction, due to a rapidly expanding thyroid lymphoma, which required emergency intervention. Our patient was submitted to a total thyroidectomy based on her symptoms since we did not have a definitive diagnosis at the time, Fine Needle Aspiration Cytology (FNAC) was only suggestive of a lymphoproliferative disorder. Six sessions of rituximab, cyclophosphamide, hydroxydaunorubicin hydrochloride, vincristine, and prednisone (R-CHOP) were indicated after a post-operative histopathology report confirmed the diagnosis of diffuse large B-cell lymphoma, which resulted in complete disease remission. This report indicates that even though chemo-radiation is considered the gold standard for the treatment of lymphomas, the existence of obstructive symptoms warrants thyroidectomy, as was done in this case.

### Keywords

Thyroidectomy, Lymphoma, Stridor, Emergency

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## Introduction

A thyroid enlargement, causing acute respiratory failure, due to thyroid compression or invasion of the tracheal lumen, presents as a rare clinical emergency warranting urgent surgical intervention. Thus, in these exceptional cases, total thyroidectomy, which is otherwise a routine elective operation, has to be resorted to on an emergency basis for life-threatening airway obstruction [1].

This airway compression may be caused by giant goiters (especially if extending substernal), acute intra-thyroidal hemorrhage, and thyroid malignancies (mostly due to anaplastic carcinoma, secondaries, and as in this case, a primary thyroid lymphoma) [1]. Dyspnea may also become critical in the latter group due to bilateral recurrent laryngeal nerve involvement and result in vocal cord palsy with stridor [1]. Massive bleeding severe enough to cause acute airway distress is rare and usually occurs as a result of neck trauma rather than as a result of thyroid illness [1]. The purpose of this paper is to describe one such patient who was successfully treated with total thyroidectomy in the emergency room due to ingravescant dyspnea and hypoxia, as well as to examine related data from the literature.

## Case Report

A 48-year-old female presented to the outpatient department (ENT) of HIT hospital, Taxila Cantt with acute stridor and labored breathing, suffered acute respiratory failure and passed out in the waiting area. The episode could have been fatal if not for the intervention. The patient was immediately shifted to the operation theatre. Emergency intubation was done, followed by total thyroidectomy. Tracheostomy was also performed later on once there was no hindrance by the thyroid mass.

A brief history: Eight months before her surgery, in February 2021, the patient experienced pain in the neck associated with diffuse swelling, which was hardly noticeable at that time. As per instructions of a general practitioner, the patient was recommended thyroid function tests. A diagnosis of hypothyroidism was made and thyroxine was prescribed. To the patient's dismay, there was no improvement in pain; instead, she developed breathing difficulties, which intensified with time. In the first week of October 2021, there was an unanticipated increase in the size of the swelling, and within two weeks it presented with limited neck extension, and severely restricted neck flexion. Meanwhile, orthopnea was also initiated, which was relieved with semi-recumbent positioning to help her sleep. Just four days from surgery, the patient presented with extreme dyspnea and a salbutamol inhaler was advised, which was not complied with. A fine needle aspiration was performed with results of no diagnostic relevance reported. The dyspnea worsened and she presented to the Heavy Industries Taxila Hospital on 27th October 2021.

Management procedure: On laryngoscopy, the laryngeal opening was compressed by the thyroid mass and was deviated to the left. Emergency intubation was done with a Rapid Sequence Induction Technique (RSII); a reinforced endotracheal tube was passed and checked for bilateral air entry. The surgical team opted for an endotracheal intubation. The intubation though riddled with complications was achieved without any major

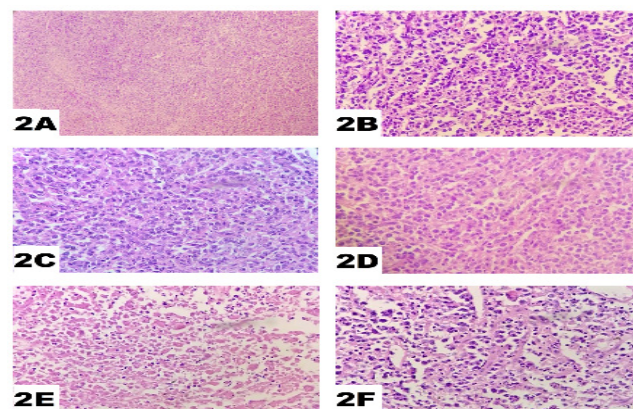
complications.

The initial surgical plan indicated an isthmectomy, which was later replaced by a total thyroidectomy when the surgical exploration revealed a large thyroid tumor involving both the lobes and extending down to the retrosternal and parapharyngeal spaces. There was bilateral involvement of lymph nodes, at the levels IIA, III, IV and V. 420 grams of thyroid tissue were removed and a tracheostomy was performed to relieve breathing. Unfortunately, intraoperatively left recurrent laryngeal nerve was damaged, which resulted in the development of hoarseness of voice. The patient underwent 6 cycles of R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone).

Histopathological findings suggested classical features of Non-Hodgkin's lymphoma. This was a primary lymphoma of the thyroid gland, though lymphomas in this anatomical sector can be of extra-thyroid origin too. Vesicular nuclei, showing a high nucleocytoplasmic ratio (Figure 2C- red) with enhanced hematoxylin staining were conclusive of neoplasia. Apparent mitotic figures (figure 2D- blue), indicative of enhanced cell



**Figure 1.** Cut-section of the thyroid gland (post-operative) with visible areas of central necrosis; the gross morphology is suggestive of a thyroid neoplasm



**Figure 2.** Figure 2A(10X), 2B(40X): Hematoxylin and eosin staining shows a diffuse monomorphic population of large atypical cells. Figure 2C, 2D (40X): Vesicular nuclei can be observed in tumor cells, having prominent nucleoli and mitotic figures Figure 2E (40X): Focal areas of necrosis are visible as preserved cell outlines without nuclei. Figure 2F (40X): Sclerotic bands surrounding groups of neoplastic cells can also be seen

division also suggested similar findings regarding neoplasia. Histopathological analysis indicated a diffuse Large B-cell lymphoma, Post Germinal type, with strong positive expression of CD 20 and MUM 1, it was also weakly positive for BCL 2. There was a negative expression of TTF 1, PAX 8, CD 3, CD 10, BCL 6 and C-MYC.

## Discussion

Despite the fact that Hedenus [1] recorded thyroidectomy for “suffocating” goitres in 1821, airway obstruction due to goitre is now uncommonly mentioned in the literature due to improved diagnostic methods and earlier treatment. Although this spectacular occurrence appears to be more common in underdeveloped countries due to ignorance and lack of inexpensive medical treatments, the phenomenon of enormous goitres is rarely reported in Western countries as well [1]. However, since the tracheal lumen can be gradually constricted without presenting symptoms up to 75% of the cases, truly severe life-threatening airway obstruction is now an exceptionally unusual occurrence [1].

Spontaneous intrathyroidal hemorrhage, invasion of the tracheal lumen by primitive or secondary tumors, severe compression from benign or malignant masses, and bilateral vocal cord palsy resulting from infiltration of recurrent nerves from thyroid malignancy are the four etiopathogeneses of severe respiratory distress in non-traumatic thyroid disease [1]. Our patient is known to have hypothyroidism as per history, which coincides with Hashimoto’s thyroiditis. Hashimoto is a potentially benign condition but it is the cause of many malignancies, especially lymphomas derived from MALT. In individuals who arrive with an increasing neck mass and a history of Hashimoto’s thyroiditis, it is imperative to rule out primary thyroid lymphoma since Hashimoto’s is the most significant risk factor for PTL [2], implying that Hashimoto’s disease is part of an immunological response that elicits primary thyroid lymphoma. Regardless of the underlying pathology, it is strongly indicated that prompt treatment must ensue once goiter is associated with features of airway compression, including dyspnea, orthopnea and stridor. Once significant respiratory compromise sets in, it is recommended to hospitalize such patients aimed at close observation, prompt workup and the need for imminent intervention. The size of a goitre is a poor predictor of upper airway obstruction when measured. Consequently, CT scanning has become the gold standard for deciding the extent of retrosternal goitre and its relationship to neighboring structures, and it can be used to identify patients who might need a thoracic intervention [3]. It is noteworthy that asymptomatic multinodular goiter can unpredictably result in airway obstruction, regardless of the duration, volume, or intrathoracic extension of the goiter. In these patients with upper airway compression, obstructive findings on pulmonary function test also do not relate with tracheal reduction on CT scans. Recognition of these cases is imperative, as they constitute a preventable cause of mortality if timely diagnosed and treated. Thyroid lymphomas, the cause of airway compromise in our case, represent a small but significant subset of such patients. Primary thyroid lymphomas represent 5% of thyroid malignancies and 2% of extra-nodal

lymphomas typically present as progressively enlarging goiters, sometimes in the backdrop of Hashimoto’s thyroiditis, which may present as diagnostic difficulty because of the mostly nonspecific sonographic features and low diagnostic yield of FNAC in the absence of Immunophenotypic analysis. Diffuse Large B-cell lymphomas and more indolent MALT lymphomas variety comprise the most common primary thyroid lymphomas. In terms of the histopathology, diffuse Large B-Cell Lymphoma (DLBCL) is classified into two types as follows: 1) Germinal Center DLBCL and 2) Post-Germinal Center DLBCL/Activated B-cell like lymphoma. CD10, BCL6 and MUM-1 immunohistochemistry markers are directive for diffuse large B-Cell lymphoma (DLBCL). According to the Hans Algorithm [4], to approximate molecular subtypes based on Immunohistochemical patterns; positive findings of CD10 or BCL6 are conclusive of the Germinal Centre (GC) type. If MUM-1 is positive then it is classified as Post-Germinal Centre (Post-GC). Positive expression of CD20 is specific for a B cell Lymphoma because it is the baseline immunofluorescence marker for B cells. GC DLBCL has a better prognosis than the Post GC DLBCL with CHOP therapy with an overall survival of 60% [5]. When activated B cell DLBCL is treated with normal chemoimmunotherapy, the outcomes are significantly worse. [6]. Immunostaining, like cDNA microarrays, can be used to distinguish between GCB and non-GCB subtypes of DLBCL and predict their survival.

Radiotherapy for local disease, and Combined Modality with chemotherapy, typically involving R-CHOP (Rituximab, Cyclophosphamide, Vincristine, Doxorubicin and Prednisolone) form the mainstay of treatment of stage III E and IV PTL. Chemotherapy with or without radiation is the indicated treatment option. These noninvasive methods offer a considerable mortality benefit over surgical resection, which is generally not recommended as first-line therapy [7].

Establishing a secure airway and induction of general anesthesia presents the first challenge when the patient in such a dramatic event presents to the emergency or operation suite. Awake fiberoptic intubation using a small endotracheal tube followed by induction of general anesthesia may offer a quick and safer approach compared to a standard sequence of induction and intubation but the later approach may be preferred in unsafe patient. On the other hand, an inhalation induction followed by laryngoscopy and orotracheal or blind nasal intubations, may be considered dangerous because of the complete airway obstruction following loss of consciousness [1].

The second step is the choice of surgical treatment to be performed. Indeed, emergency surgery is always indicated for severe airway obstruction caused by a thyroid mass [1]. In the presence of thyroid mass, an emergency tracheostomy is hindered, while an isthmectomy to allow a tracheostomy appears to be an incomplete treatment. Total, near-total or sub-total thyroidectomy represents the treatment of choice of acute airway obstruction resulting from compression of thyroid mass. Lastly, after establishing a definitive airway via endotracheal intubation, the role of tracheostomy at the end of the procedure is questionable, as a total thyroidectomy is capable of resolving airway obstruction, thus tracheostomy may be deemed unnecessary. However, it can still be justified to perform tracheostomy if there is marked tracheomalacia

[1]. There is a higher risk of postoperative hypoparathyroidism, vocal cord palsy and hemorrhage reported for thyroidectomies performed for acute airway obstruction and retrosternal goiters. Continuous intraoperative nerve monitoring is a safe and effective way to minimize Recurrent laryngeal nerve paralysis in thyroid surgery. [8].

#### **Conclusion**

In conclusion, on the basis of our experience and of the literature review, we strongly advocate elective surgery for patients with thyroid disease at the first signs of tracheal compression as the first modality of intervention in obstructing thyroid lymphomas; once a patent airway is maintained, chemotherapy can follow later on the treatment timeline. When acute airway distress appears, an emergency life-threatening total thyroidectomy is recommended in a high-volume center.

#### **Scientific Responsibility Statement**

*The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.*

#### **Animal and human rights statement**

*All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.*

#### **Conflict of interest**

*None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.*

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